**Introduction:** Congenital left coronary ostial atresia or severe stenosis is an extremely rare coronary abnormality. The clinical picture is either cardiac failure in the small infant or chest pain in the older child or adult patient. This report describes four neonatal cases of this rare abnormality.

**Methods:** We retrospectively accessed all cases of left coronary ostial stenosis or atresia seen in our center during an 11-year-period (2000 – 2011). Two older patients with this diagnosis seen at 18 months and 4 years were excluded from the study.

**Results:** Four neonates with the diagnosis of left coronary ostial atresia or stenosis were identified during the study period. One newborn died within minutes, the other within hours after birth because of cardiac failure refractory to all treatment strategies. In both cases left coronary stenosis (one case with a "pinpoint" orifice and the other with a "slitlike" orifice) was diagnosed at autopsy. The third neonate was in cardiac failure due to a severe aortic stenosis. Left coronary ostial atresia was diagnosed during emergency catheter procedure. The infant subsequently died after the aortic dilatation. The forth infant had a cardiac arrest at the third day of life after normal delivery and neonatal adaptation. She was diagnosed with left coronary ostium atresia by coronary angiography performed because of persistent biventricular dysfunction. She died during the attempt of revascularization surgery at 2 weeks of life.

**Conclusion:** Congenital left coronary ostium atresia or stenosis is very rare. Coronary angiography is the diagnostic method of choice especially in the small child or infant. Revascularization surgery seems indicated in symptomatic children based on case reports and smalls series. The clinical picture described here for the first time in the neonate is dramatic and quickly fatal with scarce surgical options. Systematic examination of the coronaries should be part of any neonatal autopsy.

## 279

### The transcatheter closure of Atrial Septal Defect (ASD) in patients older than 60 years : Retrospective study of 41 cases

Aimé Bama, Claire Dauphin, Jean Paul Chadefaux, Jean René Lusson
*CHU Gabriel Montpied, cardiologie A, Clermont Ferrand, France*

**Introduction:** Transcatheter closure of ASD is now proposed as first line treatment for the elderly patients.

**Purpose:** The aim of our study is to evaluating the transcatheter closure of ASD in patients older than 60 years.

**Methods:**The records of 41 patients (37 women, 4 men) who are over 60 years (69.88±6 years, range 61-82 years), referred between April 1998 and December 2010 for transcatheter closure of secundum ASD, were retrospectively reviewed

**Results:** ASD was discovered during the assessment of patients with dyspnea (17) or arrhythmia (17). Average age at time of diagnosis was 62.4±14.89 years (range 14-82 years). 37.8% of cases had a delayed diagnosis.

At time of the procedure, 19 patients (46.3%) had supraventricular arrhythmia and 82.9% of patients were dyspneic. Shunt ratio was 2.50±0.66, defect size 19.67±6.76 mm, stretched diameter 24.36 mm±5.87, device diameter of 24.49 mm±6.58. One patient was recused, because of increased left atrial pressure at occlusion test.

Fluoroscopy time was 7.01±4.41 min.Success rate is 97.5%. Complete closure rate at one day and one month was 84.6% and 94.9% respectively. Systolic pulmonary pressure decreased from 49.37 to 34,58 mmHg at one month.

Hospital stay was 3.58 days ± 2.71.Periprocedural complication rate was 12.2%. 2 complications were major: one death by esophageal hematoma, and one oropharyngeal bleeding, and 3 minors: 3 femoral arteriovenous fistulas.

At late follow-up (34±44 months, range 1month to 12 years), 34 patients were in stage one, 4 recurred supraventricular arrhythmia, 1 needed pace maker for sinus node dysfunction.

**Conclusion:** Transcatheter closure of ASD in ederly patients is effective. It does not change rhythmic status but allows symptomatic improvement.

## 280

### Circulating endothelial cell levels decrease after vasodilator therapy and are a biomarker of deterioration in pediatric pulmonary hypertension

Marilyne Lévy (1), Damien Bonnet (1), Laetitia Mauge (2), Clément D'Audigier (2), Isabelle Szezepanski (1), Pascale Gaussem (2), David Smadja (2)
*(1) Hôpital Necker, cardiologie congénitale et pédiatrique, Paris, France – (2) Inserm Unité 765, Faculté de Pharmacie, AP-HP, Hôpital Européen Georges Pompidou, Paris, France*

**Background:** Pulmonary vasodilators in general and prostacyclin therapy in particular have markedly improved the outcome of patients with pulmonary arterial hypertension (PAH). Endothelial dysfunction is a key feature of PAH and we previously described that circulating endothelial cells (CECs) could be used as a biomarker of endothelial dysfunction in PAH. We now hypothesized that PAH-specific vasodilator therapy might decrease CEC numbers.

**Methods:** We quantified CECs in peripheral blood from children with idiopathic PAH (iPAH, n =30) or PAH secondary to congenital heart disease (PAH-CHD, n =30), before and after treatment and during follow up. CEC were enumerated by immunomagnetic separation with mAb CD146-coated beads.

**Results:** CEC counts were significantly decreased in children after treatment with oral endothelin antagonists and/or PDE5 inhibitors. In 10 children with refractory PAH despite combination oral therapies, SC treprostinil was added and we found a further significant decrease in CEC count during the first month of treatment in every patient. We quantified CEC during 6 to 36 months follow-up after initiation of SC treprostinil and found that CEC count is modified according to clinical status.

**Conclusions:** CEC counts fall with vasodilatator therapy in PAH and could also be used as biomarker of deterioration in refractory pediatric pulmonary hypertension treated with SC treprostinil.

## 281

### Use of covered stents for closure of fenestration in extracardiac cavopulmonary connection: technical aspects and mid-term results

Sophie Guiti Malekzadeh Milani, Antoine Legendre, Damien Bonnet, Younes Boudjemline
*Necker EM, cardiologie pédiatrique, Paris, France*

**Objective:** Closure of fenestration in a cavopulmonary connection (TCPC) is performed with devices used to close intracardiac or aorto-pulmonary connections. This study presents our data regarding the use of covered stents in patients with fenestrated TCPC.

**Methods:** We retrospectively reviewed the data of all the patients receiving a covered stent to close a fenestration of TCPC between 2005 and 2012.

**Results:** 38 patients were included. Mean age and weight were respectively 7.6 years and 20 kg. Femoral access was mainly used (32) but also jugular access (5) and trans-hepatic access (1). A Cheatham Platinum Stent was used (34) but we also used Atrium V12 Stents (4). The balloon size was chosen according to the diameter of the conduit. We used BIB balloon (14) or simple balloon (Tyshak or Balt) (16) (balloon not reported (4)). Mean procedural and fluoroscopy times were respectively 42,5 +/ – 21 and 7,5±6,6 minutes. Mean central venous pressure rose from 10 to 12mmHg. Mean oxygen saturation increased from 90% to 96%. There was one anaphylactic shock at the end of the procedure and one air embolisation without hemodynamic compromise. Full occlusion was confirmed on angiogram in 36 patients. 2 cases required 2 stents to achieve full occlusion. There was one minimal residual shunt and one failure of the procedure with significant residual shunt in a patient with a conduit made of a Gore-Tex patch between the atrium and the pulmonary arteries. There were no embolic event, acute venous thrombosis or arrhythmias. Patients were treated with anti vitamin K for 6 months then aspirin. Mean follow-up is 49 months without thrombo-embolic complication or desaturation.

**Conclusion:** Covered stent is an option to close fenestration in extracardiac TCPC. It is safe, easily achievable with low fluoroscopy time, very low risk of thrombo-embolic events or failure. The good results are sustainable. The main advantage of the technique is to avoid prosthetic device in the atrium